TOURETTE'S DISORDER

Introduction

Tourette's disorder is an inherited neurological disorder characterized by repeated involuntary motor and vocal tics (Murphy et al., 2001). A tic is defined as a sudden, quick, recurrent, nonrhythmic motor movement or vocalization (Murphy et al.). Tourette's disorder is related to age and is more prevalent in children than adults (American Psychiatric Association, 2000). The diagnosis of Tourette's disorder is generally made before the child's eighteenth birthday (American Psychiatric Association). The average onset age of Tourette's disorder is between seven and ten years (National Institute of Neurological Disorders and Stroke [NINDS], 2007). Tourette's disorder affects all ethnic groups but males are three to four times more often affected than females (National Institute of Neurological Disorders and Stroke). With time, tics become more frequent and increase in variety, involving more body parts such as the trunk or legs, and often become more disruptive to activities of daily living (Medical Center of Central Georgia, 2002).

Tics can occur in any part of the body (American Academy of Child & Adolescent Psychiatry [AACAP], 2000). Chronic tics are the most prominent feature of Tourette's disorder (Kurlan, 2002). Simple vocal tics include chronic sniffing, grunting, throat clearing, clicking and screaming (Brody, 2005). Complex vocal tics can include speech interruptions such as stuttering and repetition (Brody). Simple motor tics may include eye-blinking, nose wrinkling, jaw thrusting, shoulder shrugging, or neck jerking (Brody). More complex motor tics may take the form of jumping, touching, twirling when walking, retracing steps, imitating someone else's movements, or making sudden obscene gestures (Brody). Expression of tics occurs in bouts that can be separated by seconds, hours, weeks or even months (Brody). Although tics do not go away during sleep, they are often significantly diminished (NINDS, 2007). Much like an itch, the tic provides the child with temporary relief (Brody).

Tourette's manifests itself differently in males and females. Males are more likely to have chronic tics or full-blown Tourette's, while females are more likely to have obsessive-compulsive symptoms (Ohio State University Medical Center, 2005). In all patients diagnosed with Tourette's disorder, sudden, explosive outbursts of behavior are reported in approximately 25% of patients, with such outbursts occurring more frequently in children than in adults (Budman et al., 2000). Such volatile outbursts in children with Tourette's disorder are usually accompanied by feelings of mounting tension and spontaneous activation (Budman et al.). Table 1 outlines basic facts about Tourette's disorder.

Table 1

Facts about Tourette's Disorder

- Tourette's disorder is a tic disorder.
- It is a rare disorder found more commonly in males.
- When diagnosing Tourette's disorder, Wilson's and Huntington's diseases must be ruled out.

Source: Murphy et al., 2001.

Usually, facial tics, such as rapid eye blinking or twitches of the mouth, are the first indication to parents that their child may have Tourette's disorder (the National Alliance for the Mentally Ill [NAMI], 2002). In other children, tics of the limbs or involuntary sounds, such as throat clearing and sniffing, may be initial signs. Furthermore, vocal tic activity usually involves loud grunting, but may also include word shouting, with the words sometimes being obscenities. This is called coprolalia (Murphy et al., 2001). However, only 15% of all patients diagnosed with Tourette's disorder manifest this symptom (Tourette Syndrome Association, 2002). Complex vocal tics can also include echolalia, a repeating of words or phrases of others (NINDS, 2007). Table 2 lists the categories of tics.

Table 2

Categories of Tics

Simple

Motor—Eye blinking, head jerking, shoulder shrugging and facial grimacing

Vocal—Throat clearing, yelping and other noises, sniffing and tongue clicking

Complex

Motor—Jumping, touching other people or things, smelling, twirling about, and only rarely, self-injurious actions including hitting or biting oneself

Vocal—Uttering words or phrases out of context and coprolalia (vocalizing socially unacceptable words)

Source: Tourette Syndrome Association, Inc., 2002.

The natural course of Tourette's disorder varies and, although Tourette's disorder symptoms can be very mild to quite severe, the majority of cases fall in the mild category (NINDS, 1995). Most children experience their worst symptoms of Tourette's disorder during their early teens (NINDS).

Tourette's disorder is a variable expressive disorder, which means that the Tourette's gene will result in differences in expression for different people (Ohio State University Medical Center, 2005). A recent study suggests that potentially 750,000 children in the United States have Tourette's disorder (Brody, 2005).

Diagnosis

An evaluation of the child's family history, along with general observation of the symptoms, is the most common method for diagnosing Tourette's disorder. However, before a diagnosis of Tourette's disorder is made, both motor and vocal tics must have been present for at least one year (NINDS, 1995). Neuroimaging studies may be used to rule out other conditions that might be confused with Tourette's disorder, but there are no specific laboratory tests that definitively diagnose the disorder (NINDS).

Causes and Risk Factors

Tourette's disorder is highly hereditary, with evidence supportive of genetic transmission (Murphy et al., 2001). Scientists have identified a rare gene mutation that could contribute to Tourette's syndrome. A study conducted by Yale University focused on a gene called SLITRK1. A glitch in that gene was observed. Preliminary data supports that this gene plays a role in Tourette's syndrome (Hitti, 2005).

Studies have shown that Tourette's disorder is an autosomal dominant disorder. This means that both males and females are affected, and one copy of the gene is necessary to have the condition (Medical Center of Central Georgia, 2002). A parent with Tourette's disorder has a 50% chance of passing the gene to a child (NAMI, 2002). However, a non-genetic cause for Tourette's disorder may cause up to 10 to 15% of children diagnosed with the disorder (Ohio State University Medical Center, 2005). Complications of pregnancy, low birth weight, head trauma, carbon monoxide poisoning, and encephalitis are thought to be associated with the onset of non-genetic Tourette's disorder (Medical Center of Central Georgia).

Comorbidity

According to NAMI, 40% of children and adolescents who have Tourette's disorder also have attention problems (2002). Thirty percent have academic difficulties. In fact, it is thought that approximately 50% of children with Tourette's disorder meet criteria for attention deficit hyperactivity disorder (ADHD). Most have normal intelligence and do not usually have primary learning disabilities. Some–25 to 30%–also experience symptoms of obsessive-compulsive disorder or have other forms of anxiety. Learning disabilities are common, as well as developmental stuttering. Social discomfort, self-consciousness, and depressed mood frequently occur, especially as children reach adolescence. Adolescents with Tourette's disorder may also display a variety of psychopathological conditions, such as depression, anxiety, and conduct disorder (Kurlan, 2002). Certain personality traits like irritability, argumentativeness, stubbornness and impulsivity may also represent the disorder (Kurlan).

Promising Treatments

There is no standard treatment modality for Tourette's disorder (Christophersen & Mortweet, 2001). Because manifestations of Tourette's disorder can be quite variable, children should be evaluated with great care in order to determine which aspects of the disorder are most disabling. For most children, this can serve as a guide to target specific treatment interventions.

The development of a child diagnosed with Tourette's disorder may proceed normally, with no need for treatment (Medical Center of Central Georgia, 2002). However, if tics interfere with functioning, school performance, or other disorders present, treatment may be necessary. Children with Tourette's disorder can generally function well at home and in school. If they have

accompanying emotional or learning problems, they may require special classes, psychotherapy, and/or medication (Medical Center of Central Georgia).

When symptoms interfere with functioning, medication can effectively improve attention span, decrease impulsivity, hyperactivity, tics, and obsessive-compulsive symptoms. However, behavioral interventions may also be useful for tics and symptoms associated with any co-occurring disorders (NAMI, 2002). Table 3 outlines treatment considerations for Tourette's Disorder.

Table 3

Treatment for Tourette's Disorder

Specific treatment for Tourette's disorder should be based on: • age, overall health, and medical history, • severity of tic behavior, • tolerance for specific medications, procedures, and therapies, • predictions for course of the disorder, • personal opinion and preference.

Source: Ohio State University Medical Center, 2005.

Behavior Treatments

Positive reinforcement programs appear to be most helpful in the management of tic disorders (Bagheri, 1999). Goals for target behaviors may be categorized into two groups: (1) skill deficiencies, or areas that initially require concentration to build social and academic skills; and (2) behavior excesses, in which the goal is to help the patient decrease the frequency of these behaviors (Bagheri). It is imperative that caution is employed in the management of behavior excesses, since some children who undergo behavior modification to target the Tourette's symptoms have an exacerbation of symptoms (Bagheri). The following is a brief description of treatments for the behaviors associated with Tourette's disorder.

> *Habit covariance* – refers to behaviors that, although different, frequently occur together. When one behavior changes, the other will as well. In children with Tourette's disorder, behavior treatments can prove effective for eliminating problem behaviors. However, all behaviors must be evaluated in term of age-appropriateness and properly assessed as not being appropriate for the child's age and relating to the disorder. Treatment of habit disorders must be implemented by a service provider with adequate training in order to be effective.
>
> *Habit reversal* – treats symptoms associated with Tourette's disorder. The complete habit reversal training package involves awareness training, self-monitoring of tics, relation training, competing response training, and motivational techniques (Himle et al., 2006). Habit reversal emphasizes awareness, motivation, correction and prevention and is generally well tolerated. Treatment of habit disorders must be implemented by a service provider with adequate training in order to be effective.
>
> Source: Christophersen & Mortweet, 2001.

Another treatment that has shown some promise is cognitive behavioral therapy (CBT). CBT is used to challenge and restructure the way participants assess their expectations and actions in situations that may cause frequent ticking behavior (Cook & Blacher, 2007). One study conducted to measure the effectiveness of CBT in reducing tics revealed that CBT produced reductions in tics similar to results produced by habit reversal (Cook & Blacher). While this study had certain limitations, CBT was observed as showing some promise in the treatment of Tourette's disorder.

Additional research is being conducted on the effectiveness of CBT in the treatment of Tourette's disorder.

Pharmacological Treatment

Medication therapy can be utilized if the symptoms of Tourette's disorder are not amenable to non-drug interventions. Medication should be chosen based on the specific symptoms, as well as potential side effects of the medication. For example, in one patient, treatment of the tic may be the goal, while treatment of obsessive-compulsive features may take precedence in another (Kurlin, 2002). Dosages should be adjusted to the lowest appropriate level.

Most children with Tourette's syndrome require medication for up to one to two years, with 15% requiring long-term medication for tic control (Bagheri, 1999). When tics appear to be controlled for a long period, a slow and gradual reduction in medication should follow (Bagheri).

New research is being conducted to determine whether a mixed dopamine agent is safe in treating children with Tourette's disorder (Gilbert et al., 2003). Neuroleptics, which block dopamine transmission, are used to treat children with severe tics (Gilbert et al.). However, major side effects resulted from this treatment (Gilbert et al.). Preliminary results of this study of mixed dopamine agents suggests a potential benefit for children with chronic tic disorders and Tourette's disorder (Gilbert et al.).

As noted by Bagheri (1999), many patients with Tourette's syndrome have comorbid conditions and treatment for these conditions may be necessary. Treatment of comorbid ADHD has been controversial because of reports that stimulants hasten the onset or increase the severity of tics in some patients. However, stimulants alone may not substantially worsen the severity of the disorder and it may prove necessary to treat both the ADHD and the Tourette's syndrome with a stimulant in combination with either clonidine or guanfacine, or with a neuroleptic agent. However, the use of several drugs or medicines together in the treatment of Tourette's disorder should be minimized, especially in children (Bagheri). Table 4 shows the pharmacotherapy currently used to address the symptoms associated with Tourette's disorder.

Furthermore, according to Bagheri (1999), the treatment of the co-occurring obsessive-compulsive disorder with selective serotonin reuptake inhibitors (SSRIs) may prove effective. However, there is often a delay between commencement of medication and the intended pharmacological response. Moreover, this response may take as long as four to six weeks (Bagheri). Behavior therapy may also be used in treating the co-occurring disorder of obsessive-compulsive disorder.

Unproven Treatments

Research has shown the lack of evidence to support several treatments for Tourette's disorder. One such treatment is plasma exchange or intravenous immunoglobulin (IVIG), treatment. In fact, the National Institute of Mental Health (NIMH) and the Tourette Syndrome Association have advised that there is no evidence of their efficacy in children with Tourette's disorder and both treatments carry a potential for significant adverse reactions (NIMH, 2000).

Massed negative practice is based on the premise that over-rehearsal of the tic by the patient leads to its disappearance (Cook & Blacher, 2007). However, studies have shown that massed negative practice has failed to produce reductions in tics comparable to reductions produced by

habit reversal (Cook & Blacher). There are also contradictory studies regarding the effectiveness of contingency management to reduce tic frequency (Cook & Blacher).

Table 4

Pharmacotherapy of Tourette's Disorder

Tics	
Neuroleptics	Clonidine
Haloperidol	Other Drugs
Pimozide	Botulinum Toxin*
Fluphenazine	
Others	
Obsessive-Compulsive Disorder	
Clomipramine	Sertraline
Fluoxetine	
Attention Deficit Hyperactivity Disorder	
Clonidine	Stimulants
Tricyclic antidepressants	Methylphenidate
	Pemoline
	Dextroamphetamine

*Recent research has shown that, for a small number of patients who prove resistant to the motor medications, injections of botulinum toxin might be helpful.

Source: Kurlan, R., 2002.

Recent studies on treatment for Tourette's disorder describe attempts to relieve symptoms of the disease through deep brain stimulation (Brody, 2005). This type of treatment involves implanting electrodes in the brain where movement is controlled (Brody). This type of treatment is still highly experimental, with no data on its overall effectiveness, potential complications, side effects, or duration of benefit (Brody).

Another new treatment approach involves temporarily paralyzing the affected muscle group with a botulinum toxin, which has the potential to suppress the tic for several months (Brody, 2005).

Other Important Treatment Elements

It is important to realize that simple inattention or hyperactivity by itself is not sufficient for diagnosis.

Cultural Considerations

Tourette's disorder is universally prevalent. However, the understanding of the disorder varies significantly in that tic symptoms are not considered a problem and are not usually mentioned to physicians (Mathews, 2001). Many families may consider the tics to be a bad habit, and health care professionals, when consulted, may concur. In Latin American countries, such as Costa Rica, tics and obsessive symptoms presented by children with Tourette's disorder may be considered annoying and perhaps unattractive but not otherwise noticed (Mathews). Tics may even be thought to be voluntary in nature.

For example, symptoms that would be reported as causing significant impairment in children in the United States were often reported as having little or no impact, primarily because the needs and

expectations of these cultures were different (Mathews, 2001). Studies reveal that, because concepts such as impairment can be culturally defined, specified diagnostic criteria are not always adequate for purposes of identifying Tourette's disorder as a true mental health disorder. Such views certainly influence diagnosis and treatment.

Sources

American Academy of Child & Adolescent Psychiatry (AACAP). (2000). *Tic Disorders*. [Online]. Available: http://www.aacap.org/publications/factsfam/tics.htm. [March 2005].

American Psychiatric Association. (2000). *Diagnostic and Statistical Manual of Mental Disorders*, *Fourth Edition, Text Revision.* Washington, DC. American Psychiatric Association.

Bagheri, M. (1999). *American Family Physician*. Recognition and Management of Tourette's Syndrome and Tic Disorders**.**

Brody, J. (2005). The Tics of Tourette's Often Go Undiagnosed. *The New York Times.* [Online]. Available: http://www.nytimes.com/2005/01/18/health/18brod.html. [January 2005].

Bruun, R., Cohen, D., & Leckman, J. (1999). *Guide to the Diagnosis and Treatment of Tourette Syndrome and Other Disorders.*

Budman, C., Bruun, R., Park, K., Lesser, M., & Olson, M. (2000). Explosive Outbursts in Children with Tourette's Disorder. *Journal of American Academy of Child & Adolescent Psychiatry, 39* (1270).

Christophersen, E., & Mortweet, S. (2001). *Treatments That Work With Children: Empirically Supported Strategies for Managing Childhood Problems.* American Psychological Association.

Cook, C. & Blacher, J. (2007). Evidence-based Psychosocial Treatments for Tic Disorders. *Clinical Psychology Scientific Practices, 14*, 252-267.

Gilbert, D., Dure, L., Sethuraman, G., Raab, D., Lane, J., & Sallee, F. (2003). Tic Reduction with Pergolide in a Randomized Controlled trial in Children. *Neurology*. [Online]. Available: http://www.neurology.org/cgi/content/abstract/60/4/606. [March 2005].

Himle, M., Woods, D., Piacentini, J., & Walkup, J. (2006). Brief Review of Habit Reversal Training for Tourette Syndrome. *Journal of Child Neurology*, *21*, 719-725.

Kurlan, R. (2002). Current Pharmacology of Tourette Syndrome. *Tourette Syndrome Association, Inc*. [Online]. Available: http://www.tsa-usa.org. [October 2002].

Mathews, C. (2001). Cultural Influences on Diagnosis and Perception of Tourette Syndrome in Costa Rica. *Journal of the American Academy of Child & Adolescent Psychiatry*. April, 2001.

Medical Center of Central Georgia. Child and Adolescent Mental Health. (2002). *What is Tourette's Disorder*? [Online]. Available: http://www.mccg.org/childrenshealth/mentalhealth/tourette.asp. [October 2002]. *Not available July 2005*.

Murphy, M., Cowan, R., & Sederer, L. (2001). Disorders of Childhood and Adolescence. *Blueprints in Psychiatry, Second Edition*. Malden, Mass: Blackwell Science, Inc.

National Alliance for Mentally Ill (NAMI). (2002). Tourette's Syndrome Fact Sheet. [Online]. Available: http://www.nami.org/helpline/tourette.html. [October 2002].

National Institute of Mental Health (NIMH). (2000). Warning about Two Therapies for Tourette's, OCD (Obsessive-compulsive Disorder). [Online]. Available: http://intramural.nihm.nih.gov/research/pdn/web.htm. [October 2002]. *Not available July 2005.*

National Institute of Neurological Disorders and Stroke of the National Institutes of Health (NINDS). (1995). NIH Publication No. 95-2163. Tourette Syndrome. [Online]. Available: http://www.ninds.nih.gov/patients/disorder/tourette/tourette.htm. [October 2002]. *Not available July 2005*.

National Institute of Neurological Disorders and Stroke of the National Institutes of Health (NINDS). (2007). *Tourette Syndrome Fact Sheet*. [Online]. Available: http://www.ninds.nih.gov/disorders/tourette/detail_tourette.htm#96193231. [November 2007].

Ohio State University Medical Center. (2005). *Tourette's Disorder*. [Online]. Available: http://medicalcenter.osu.edu/patientcare/healthinformation/diseasesandconditions/mentalhealth/children/tourettes. [March 2005].

Hitti, M. (2005). *Gene Link to Tourette's Syndrome Found*. News Release, Yale University. WebMD Medical Reference.

Tourette Syndrome Association, Inc. (2002). *What is Tourette's Syndrome?* [Online]. Available: http://www.tsa-usa.org. [October 2002].

Organizations/Weblinks

American Academy of Family Physicians

Information from Your Family Doctor
Understanding Tics and Tourette's Syndrome
http://www.aafp.org/afp/990415ap/990415f.html

Children and Adults with Attention Deficit/Hyperactivity Disorders (CHADD)

8181 Professional Place, Suite 201 - Landover, MD 20785
National Call Center 800-233- 4050
http://www.chadd.org

National Alliance for the Mentally Ill (NAMI)

Tourette's Syndrome
http://www.nami.org/Content/ContentGroups/Illnesses/Tourette.htm

National Institute of Neurological Disorders and Stroke

http://www.ninds.nih.gov/

National Institutes of Health (NIH)
NIH Publication No. 95-2163. Tourette Syndrome.
http://www.ninds.nih.gov/disorders/tourette/detail_tourette.htm

Obsessive-Compulsive Foundation, Inc. (OCF)
90 Depot St., P.O. Box 70 - Milford, CT 06460-0070
203-878-5669
http://www.ocfoundation.org

Tourette Syndrome Association, Inc.
42-40 Bell Blvd. - Bayside, NY 11361
718-224-2999.
http://www.tsa-usa.org

Tourette Syndrome "Plus"
http://www.tourettesyndrome.net

Tourette Syndrome Association, Inc.
Greater Washington, DC Chapter (serving MD, VA, WV, and DC)
E-mail TSAGW@aol.com
877-295-2148 or 301-681-4133
http://www.tsa-usa.org

Virtual Hospital
Tourette Syndrome
http://www.vh.org/adult/patient/psychiatry/tourettesyndrome/index.html